



CONGENITAL CARDIOLOGY SOLUTIONS
(PEDIATRIC CARDIOLOGY AND ADULT CONGENITAL HEART DISEASE)

BICUSPID AORTIC VALVE IN CHILDREN: ARE WE OVERESTIMATING THE RISK?

ACC Poster Contributions
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Authors: *William T. Mahle, Patricio A. Frias, Children's Healthcare of Atlanta, Atlanta, GA*

Background: There has been heightened concern about risks associated with bicuspid aortic valve (BAV), particularly dilatation of the ascending aorta. In young adults with BAV primary cardiac events occur in approximately 1.5% of patients per year. However, potential for aortic dissection or other cardiac events in children with BAV remains largely unknown.

Methods: Analysis of a large single institutional cohort of children (<18 yrs) with isolated BAV was undertaken to determine the prevalence of significant ascending aortic dilation and risk of cardiac events. Subjects with known genetic disorders and additional lesions such as coarctation of the aorta were excluded. Subjects with critical aortic stenosis who underwent intervention in the 1st year of life were excluded. Aortic dimensions were derived from echocardiography and values plotted as Z scores. Clinical outcomes included death, aortic dissection or elective cardiac surgery.

Results: 981 children with isolated BAV were identified. The mean age at diagnosis was 8.3 ± 5.1 yrs. At time of last pediatric follow-up the median peak gradient was 18 mm Hg, 7.8% had >mild aortic insufficiency and the mean Z score for the ascending aorta was 2.31. There were 9,427 pt yrs of follow-up. Primary cardiac events occurred in 38 subjects, yielding an event rate of 0.04 per 10 patient years. Ten subjects (1.0%) underwent aortic valve surgery and one subjected had aortic root surgery. Thirty subjects (3.0%) underwent balloon dilatation of the aortic valve. There was a single case of endocarditis. There were no cardiac-related deaths and no cases of aortic dissection.

Conclusions: The incidence of primary cardiac events in children with BAV is relatively low, approximately 3x lower than in young adults, and is generally related to aortic stenosis amenable to balloon dilatation. While mild ascending aortic dilation is common in children, the clinical course is relatively benign. In this series aortic dissection did not occur--and to our knowledge there are no published reports in the literature of aortic dissection occurring in children with isolated BAV. Whether elective surgery for the dilated aorta has a role in children remains unknown.